Calcifying Cystic Odontogenic Tumor: A Case Report and Review

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Calcifying cystic odontogenic tumor (CCOT) are uncommon benign slow-growing cysts of developmental origin partly exhibiting characteristics of a neoplasm. Until date, the exact behavior of CCOT had been enigmatic and still the lesion portrays a platform to be debatable. The lesion has been reviewed and redressed from the date first reported about its diverse clinical and histopathological features. The name of the lesion itself has undergone extensive review for the same reason. We report a case of calcifying odontogenic cyst in the mandible with a review of the literature. This paper also highlights the update on nomenclature and classification.

Keywords: Cyst, Calcifying, Gorlin, Odontogenic

INTRODUCTION

Calcifying odontogenic cyst (COC) which is also known as Calcifying cystic odontogenic tumor (CCOT) is defined as a mixed radiolucent-radiopaque lesion of the jaws with features of both a cyst and a solid neoplasm; characterized microscopically by an epithelial lining showing a palisaded layer of columnar basal cells, presence keratinization, dentinoid, and calcification.1 It is a hybrid lesion of the jaw exhibiting radiolucency and radiopacity showing cystic and solid neoplastic characteristics. The cyst often resembles features of ameloblastoma and may contain ghost cells, tooth like material and calcified deposits.2

The classification and nomenclature of this lesion underwent revisions many times from the day it has been reported, due to its characteristic features and behavior. Here, we report a case of CCOT and literature review highlighting the update on nomenclature and classification.

CASE REPORT

A 52-year-old female patient reported with a swelling in the anterior segment of the mandible. The swelling is present since 2 years and remained relatively of the same size. She was asymptomatic, and her medical history was not relevant. A firm, well-circumscribed, solitary, non-tender swelling was seen in the right anterior mandibular region occupying the labial and buccal vestibule measuring about 3 cm × 2 cm in diameter and extending anteriorly till the midline and posteriorly till the first premolar (Figure 1). Submandibular lymph nodes were not palpable. Overlying mucosa is normal in color. Grade I mobility was seen with teeth #32, #31, #41, #42 and #43.

Panoramic radiograph revealed a well-circumscribed unilocular radiolucency with tiny radio-opaque flecks within it, extending from the apical region of #31-44 (Figure 2). The lesion was surrounded by a sclerotic margin. Based on the clinical and radiographical features, differential diagnosis of calcifying epithelial odontogenic tumor, ossifying fibroma and CCOT were assigned. Radiographic features suggestive of odontoma or adenomatoid odontogenic tumor were not present. Fine needle aspiration cytology was inconclusive. Incisional biopsy revealed a cystic lesion showing lining of proliferative odontogenic epithelium similar to ameloblastoma (Figure 3). Few cells are cuboidal and stellate reticular like cells are seen superficial to ameloblast like cells. Excisional biopsy showed cystic lumen with proliferating epithelium with areas of dystrophic calcifications (Figure 4). Numerous large, homogeneously eosinophilic cells of varying sizes with pyknotic nuclei suggestive of “ghost cells” were seen. Few of the ghost cells stained with hematoxylin were suggestive of ghost cell undergoing calcification (Figure 5). Adjacent to the ghost cells...
cells, irregular basophilic masses suggestive of dystrophic calcification were also seen. The connective tissue showed numerous endothelial lined blood vessels and mixed inflammatory cell infiltrate, predominantly lymphocytes. Within the capsule, some areas show eosinophilic masses suggestive of dentinoid material beneath the epithelial lining. Proliferating strands and cord of ameloblastomatous cells were not observed in the wall, ruling out the proliferative variant dentinogenic ghost cell tumour (DGCT). The lesion was diagnosed histopathologically as simple cystic, non-proliferative CCOT and it was surgically enucleated under general anesthesia. The follow-up was done till 2013 and recurrence was nil.

**DISCUSSION**

A wide span of descriptive terminologies have been assigned to address this lesion till date due to the diverse behavior of calcifying odontogenic cyst (COC). Gorlin initially called COC as Gorlin’s cyst and was termed as keratinizing and COC by Gold in 1963. Gorlin reported that COC may be the oral analogue of dermal calcifying epithelioma of Malherbe. Precedently, the first author to publish COC was Rywkind and later on he termed it as cholesteatoma of jaw.
enlist the history of nomenclature of this lesion.3,6-15 The term COC was given by World Health Organization (WHO) in 1992 and referred it a tumor like odontogenic cyst of the jaws and was categorized under benign odontogenic tumors but continued to use the term COC. COC is generally considered to be a cyst, yet many researchers choose to address it as a neoplasm, which led to sub classify the variants of the lesion into cystic and neoplastic variants. In 2005, WHO addressed the cyst as CCOT (SNOMED code 9301/0) and DGCT (SNOMED code 9302/0) representing the cystic and neoplastic variants respectively.16

The CCOT may prevail as an intrabony or extrabony form. The age of the patient may vary from first to ninth decade and thus far no gender predilection has been reported.17-19 Distribution of cases based on site for maxilla and mandible are comparable. Extrabony CCOTs most commonly occur as a swelling in the incisor-canine region calibrating up to 4 cm and are mostly asymptomatic. They usually have the appearance of well-delineated swellings having smooth surface with a pink to reddish hue.20 Extra-osseous CCOTs may reveal a shallow depression in the bone, and occasionally displacement of adjacent teeth was observed. Largely, intraosseous CCOTs are identified as unilocular radiolucencies with a well-delineated border. 50% of cases exhibit varying flecks of radiopacity. In most of the cases, this entity reveals root resorption along with root divergence and <25% of cases may show an association with an unerupted tooth.21-22

In the early stages of formation, CCOT may have little or no mineralization and therefore may present as radiolucencies. As the lesion matures, calcifications occur and appear as well-circumscribed, mixed radiolucent-radiopaque masses. Dense opacities are associated with complex odontome. Radiologically, three basic patterns of radiopacities are identified particularly salt and pepper pattern of flecks, fluffy cloudlike pattern throughout, and a “new moon”-like configuration with crescent-shaped radio-opacity on one side of the radiolucency.21 The differential diagnosis in these instances includes dentigerosous cyst, odontogenic keratocyst (OKC), and ameloblastoma. In later stages when a mixed radiolucent-radiopaque appearance is present, the differential diagnosis would include adenomatoid odontogenic tumor, a partially mineralized odontoma, calcifying epithelial odontogenic tumor and ameloblastic fibroodontoma. In the early stages of formation, differential diagnosis can be dentigerosous cyst, OKC, and ameloblastoma as they have little or no mineralization and therefore may present as radiolucencies.22

Histopathologically, CCOT reveals an odontogenic epithelium with columnar basal cells, upper layers show stellate reticulum like cells and a fibrous capsule. The lesion differs histologically from the odontogenic cysts and epithelial tumors such as ameloblastoma, but could be similar to the Pindborg’s tumor.22 Ghost cells, tooth like material and calcifications are also seen scattered in the epithelium The mechanism of formation of ghost cells is controversial; it may represent coagulative necrosis or a form of normal or aberrant keratinization of odontogenic epithelium. Some theories reveal an underlying ischemic process, which may result in squamous metaplasia and later tends to calcify.23 These eosinophilic ghost cells can be a nuclear or with pyknotic nuclei and occasionally, ghost cells tend to calcify. The dystrophic calcification of the keratin may induce a foreign body response in the fibrous capsule and may mimic pilomatrixoma of skin.20,22 Masses of ghost cells may coalesce together to form large sheets of amorphous material. Juxta-epithelially, an eosinophilic material resembling dentin may be evident owing to the inductive effect by the odontogenic epithelium on the adjacent mesenchymal tissue.24,25 However, these ghost cells are not exclusively seen in CCOT, but are also seen in other odontogenic tumors like odontoma, ameloblastic fibro-odontoma and ameloblastic odontomas.18

<table>
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<th>Table 1: Evolution of nomenclature of CCOT</th>
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<td>1962 - Gorlin et al. identified the lesion as a distinct pathological entity</td>
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<td>1963 - Gold named the lesion as KCOC</td>
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<td>1972 - Fejerskov and Krog used the term CGCOT - monistic concept</td>
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<tr>
<td>1975 - Freedman et al. introduced the term CCOT - monistic concept</td>
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<tr>
<td>1981 - Praetorius and Ledesma-Montes suggested (COC - cystic type) and (DGCT - neoplastic type) - based on the dualistic concept</td>
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<td>1986 - Ellis and Shmookler suggested EOGCT</td>
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<td>1990 - Colmenero et al. suggested OGCT</td>
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<td>1992 - WHO classification - According to Kramer and Pindborg from 1992 and the majority of the authors favored the use of the term COC and described it as a cystic or neoplastic-like odontogenic pathological lesion of the jaw and classified it as a benign odontogenic tumor</td>
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<td>1994 - Hirshberg et al. Considered COC associated with an odontoma as a separate entity and suggested the name odontocalcifying odontogenic cyst</td>
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<td>1998 - Toida named the entity as CGCOC</td>
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<td>2005 - The WHO Classification of odontogenic tumors re-named this entity as CCOT, the benign solid type was referred to as DGCT and the cases previously reported as OGCC and malignant EOGOCs were re-named by WHO as GCOC</td>
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age lesions like ameloblastoma. Absence of the associated impacted tooth and also the age of the patient ruled out adenomatoid odontogenic tumor and dentigerous cyst which fits in well with the location.

CONCLUSION

Considering the debatable nomenclature of the lesion it is advisable to conduct more studies to explore the finer details so as to propose a classification, which is universally acceptable.

REFERENCES